

OPHTHALMOPLÉGIA EXTERNA PARTIALIS.

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THE condition of paralysis of all the muscles moving the eyeball, *ophthalmoplegia externa totalis*, is well known and easily recognized. Its lesion consists of a general destructive process of a subacute inflammatory or degenerative nature in the nuclei of origin of the sixth, fourth, and third nerves upon the floor of the fourth ventricle and aqueduct of Sylvius. It is always bilateral. The destruction of the sixth nerve nucleus on either side causes a paralysis of conjugate movement of both eyes toward the side of the lesion; and when both sixth nerve nuclei are involved together, the eyes look directly inward and cannot be turned from side to side. The destruction of the fourth nerve nuclei produces a paralysis of the act of looking upward and inward; and that of the third nerve nuclei deprives the patient of all other movements of the eyeball and upper lid, so that in the terminal stage of this disease double ptosis and immobility of both eyeballs are present. The motions of the iris are, however, unaffected in this disease, and the nuclei governing the movements are not involved in the degenerative process, though they lie quite near to and just in front of those governing the other muscles supplied by the third nerve. Twenty-seven cases of this affection have been collected by Mauthner in his lectures, a number of them accompanied by autopsy; and other cases published since 1885, some of which were brought together by Mittendorf in 1887,* have established the ex-

* JOURNAL NERVOUS AND MENTAL DISEASE, 1887, p. 78.

istence of this disease and the invariable character of its lesion.

It is not to *total* ophthalmoplegia externa that I desire to call attention in this paper, but to a condition of paralysis of but one or two of the muscles moving the eyeball, a condition which may be termed *ophthalmoplegia externa partialis* to distinguish it from the condition already described.

It is of course well known that a paralysis of the external rectus muscle of one or of both eyes is a common occurrence due to a lesion affecting one or both sixth nerves in their long course upon the base of the brain. Upon this form of partial ophthalmoplegia I do not care to dwell, as it is well understood. Nor need I refer to paralysis of the patheticus or fourth nerve which occasionally occurs, but is extremely rare. It is especially to paralysis of one or two or more of the muscles governed by the third nerve that I desire to call attention. For there are several points of interest which arise in its study, and which have not been fully discussed either in neurological or ophthalmological literature.

Paralysis affecting but one or two of these muscles moving the eyeball is so rare as to warrant a report of every case observed. And the fact that six such cases with autopsies are now on record warrants an attempt at the localization of the lesion during life.

The following case has been under observation for a year, and has been in a stationary condition for the past nine months. It offers a number of points of interest for discussion.

S. D., a Frenchman, aged 56, a painter by occupation, and a resident of Providence, R. I., was brought to my clinic April 18th, 1887. He had been a healthy man all his life, with the exception of occasional attacks of rheumatism and frequent attacks of migraine. He had never contracted syphilis, and denied all symptoms of pulmonary, cardiac, gastro-intestinal and renal disease, although a physical examination revealed the existence of slight aortic obstruction producing a systolic murmur heard at the base and associated with slight ventricular hypertrophy.

He stated that about the first of April, 1887, he had been seized very suddenly with double vision and vertigo, objects appearing to move up and down constantly, so that he was much bewildered and unable to stand or to walk alone. He managed with help to reach his home, but has no recollection of what occurred during the three following days, during which, according to the statement of his family, he lay in a somnolent condition, but not comatose or paralyzed. He was then able to get up, but felt stupid, dizzy, and walked with difficulty, it being impossible for him to fix any object with his eyes; all objects being seen double and in motion. These symptoms have improved slightly, but he still feels weak, has vertigo and double vision. He has never had headache, nor has he felt any sensation of numbness or cold or pain in his body, and he has had no paralysis, tremor, or spasm.

Examination shows a well-nourished, intelligent, active man, whose facial expression is rendered peculiar by the position of his eyes. When at rest, they diverge slightly, and the right eye is turned upward, and the right pupil is slightly larger than the left. When the eyes are moved it becomes apparent that the motion is defective. The eyes can be turned from side to side together perfectly, but such motion soon produces lateral nystagmus of the right eye. They cannot be converged to an object nearer than two feet, because of slight weakness of the right internal rectus muscle. They cannot be turned downward below the horizontal line either together or when tested separately. When asked to look up, the right eye follows the object above the horizontal line, but the left eye does not. Both eyes, however, turn up and in, though this motion produces rotary nystagmus. The reaction of the pupils to light and in accommodation is prompt, though the right pupil contracts in accommodation more slowly and less completely than the other, and remains slightly larger. Tests by secondary deviation and double images confirm the conclusion reached by this examination, viz., that in the right eye there is paralysis of the inferior rectus and paresis of the internal rectus; and that in the left eye there is paralysis of the inferior rectus.

and superior rectus. There is no ptosis. There is no paralysis of the oblique muscles or of the external recti. One week later the paresis of the right internal rectus, the difficulty in convergence, and the difference in the size of the pupils had disappeared, but all the other conditions remained, and they have persisted until the present time (April, 1888). He is still suffering from general weakness, vertigo, and double vision, although the latter symptom no longer troubles him excepting when he attempts to draw lines in painting, when he finds that he does not draw accurately, and hence has had to give up his work. He often staggers in walking, but this is due to vertigo and not to ataxia, and it is not constant. He walks as well with eyes closed as with them open. Attempts to turn the eyes up or down, or upward or inward, produce nystagmus of a rotary kind, more marked in the right eye, and this always makes him dizzy. He has developed no further symptoms, is not paralyzed, has equal and normal tendon reflexes, and has no loss of sensation, vision or hearing. The diagnosis made is embolism, from the aortic valve, in the small arteries entering the posterior perforated space between the crura cerebri, and resulting in one or more small foci of softening in the tegmentum cruris.

This diagnosis can be reached by exclusion ; for it is impossible for the symptoms to have been caused by a tumor or a meningitis upon the base of the brain in the course of the third nerves. Such a lesion would not have come suddenly or have remained stationary, and would have involved the nerve as a whole, impairing all its functions and not affecting merely a part. Nor have we here a condition of acute inflammation with hæmorrhage in the floor of the aqueduct of Sylvius,* nor such a condition as occurs in a true ophthalmoplegia externa ; for there is no tendency manifest toward an extension of the symptoms, or to complete immobility of the eyes.

It is true that a hæmorrhage in the same region is with difficulty distinguished from an embolism, but in hæmorrhage some evidence of pressure upon the adjacent sensory

* Polio encephalitis of Wernicke.

or motor tracts is usually shown by unilateral symptoms, which have been wanting here; and here there is a roughened aortic valve to give rise to an embolus. The nature of the lesion is therefore easily determined.

The chief interest in this case lies in the fact that it seems to aid us in the localization of the oculo-motor nerve nuclei when taken in connection with other cases recently published, and accompanied with autopsy.

It is well known that by experimental irritation of the floor of the aqueduct of Sylvius in dogs, Hensen and Völckers claimed* to have located in rabbits the nuclei governing the various functions of the third nerve in the following order from before backward:

Ciliary muscle, accommodation.

Sphincter iridis, light reflex.

Rectus internus.

Rectus superior.

Levator palpebræ.

Rectus inferior.

Obliquus inferior.

If, however, the cases upon record of paralysis of individual muscles in combination be considered, it is evident at once that this order of the nuclei cannot be the one which obtains in man. In the case just related, for example, the rectus internus and rectus inferior of one eye, and the rectus superior and rectus inferior of the other eye, were paralyzed together, and no other muscles were affected. A single lesion would not explain the symptoms in either eye were this scheme true. It may be admitted, however, that the centres governing the movements of the iris do lie anterior to all the others. For Westphal has recently published a case in which all the nuclei governing the motions of the eyeball were destroyed, but in which two nuclei lying in the floor of the aqueduct of Sylvius near to its opening into the third ventricle were found intact. And to these nuclei he assigns the function of governing the motions of

* V. Graefe's Arch. f. Ophthal., 1887, vol. xviii., p. 1. They admit that the evidence is not wholly satisfactory.

the pupil which were preserved in his case.* And a complementary case has recently been recorded by Bernhardt, in which the action of the pupils in light and accommodation was permanently lost, with only temporary affection of the other ocular muscles on one side.† And Leube has recorded a case,‡ with autopsy, in which the action of the right pupil was suspended and the right levator palpebræ alone of all the ocular muscles was paralyzed; the lesion lying far forward beneath the corpora quadrigemina anterior. By the kindness of Dr. Seguin, I am allowed to record a case similar to these two, in which there was a paralysis of the iris for three weeks, no other muscles being at all affected.§ The conclusion from these cases with and without autopsies is that the pupillary action is controlled by centres lying independent of and farther forward than all the other third nerve nuclei.

The independence of the iris centres from those governing the ocular muscles had been reached several years ago by the investigation of Hutchinson|| upon ophthalmoplegia externa, in which it was shown that paralysis of the ocular muscles implied a progressive destruction of the various nuclei concerned in motions of the eyeball from the sixth nerve upward through the third nerve. Hutchinson supposed that a lesion in the ciliary ganglion alone produced pupillary paralysis. But this conclusion was soon shown to be erroneous, and subsequent investigations proved that when the entire gray matter of the floor of the aqueduct was involved the iris was also affected. And subsequent cases, with autopsies, have confirmed this statement. The independence of the centres of the iris from those of the eyeball does not therefore imply their wide separation.

The question remaining for settlement is the relative

* Westphal, *Arch. f. Psych.*, 1888, vol. xix., p. 858. It is true that the light reflex was lost in this case, but this W. explains by the existence of posterior spinal sclerosis.

† Bernhardt, *Berliner Gesellsch. f. Neurologie*; *Arch. f. Psych.*, 1888, xix., 505. Case 1.

‡ Leube, *Deut. Arch. f. Klin. Med.*, 1887, xl.

§ See this *Journal*, this number, page 317.

|| Hutchinson, *Medico-Chirurg. Trans.*, 1879, vol. lxii., p. 307.

order in which the remaining nuclei of the third nerve are arranged. What are the facts? There is, first, a case of Leube, (1) in which the paralysis of the pupil was accompanied by paralysis of the levator palpebræ, on the right side only. There is, secondly, a case of Bernhardt, (2) in which the paralysis of the pupil was associated with paralysis of the levator palpebræ, recti superior, inferior and internus, the latter four muscles all recovering.

There is, thirdly, a case of Thomsen,* (3) in which a temporary paralysis of both pupils and both levator palpebræ was accompanied by permanent paralysis of both superior recti.

And, lastly, there is a case of Steffen,† (4) in which double ptosis was associated with sluggish action of the pupils, both ascribed to a destruction of the corp. quadrigemina.

These cases seem to prove that the centre governing the levator palpebræ lies next in order to the centres for the iris. Let us see what muscle is most frequently affected in conjunction with the levator palpebræ.

Bernhardt‡ has recently reported another case, in which the levator palpebræ on the right side, (5) and the levator palpebræ and superior rectus on the left side, (6) were paralyzed.

Mauthner§ cites a case of v. Grafé, in which congenital abscess of the iris was accompanied by partial ptosis and imperfect elevation of the eyeballs, (7) and remarks that he has seen a similar condition without absence of the iris (8).

A year ago I published|| the history of a patient who was suddenly seized with paralysis of the left levator palpebræ and superior rectus, (9) and with slight right hemianæsthesia, all of which symptoms remained as long as the patient was under observation (five months).

In 1881, Kahler and Pick published a case,¶ with autopsy, in which the levator palpebræ and rectus superior and

*Archiv. f. Psych., xix., 185.

† Cited by Nothnagel "Topische Diagnostik," p. 214.

‡ L. c. Case 2.

§ Mauthner, Vortrage ii., 4, 370.

|| JOURNAL OF MENTAL AND NERVOUS DISEASE, 1887, p. 115.

¶ Zeitschrift f. Heilkunde, 1881.

obliquus inferior were totally paralyzed on the left side, (10) and the rectus inferior and rectus internus were weak. The lesion in this case was a hæmorrhage in the tegmentum, involving the left red nucleus and the fibres of the third nerve which pass through it, especially those in its lateral part, the median part being unaffected.

To this case they added another, (11) in which a temporary paresis of the levator palpebræ, rectus superior and inferior, was accompanied by a paralysis of the rectus internus, which persisted until death. The obliquus inferior was not affected. The autopsy showed a small hæmorrhage in the right half of the tegmentum, involving the red nucleus and the fibres of third nerve passing through its median portion.

In the case of Thomsen already cited, the temporary paralysis of the pupils and levator palpebræ was accompanied by a permanent paralysis of the superior rectus on both sides.

From these cases it seems evident that the levator palpebræ and superior rectus are so often associated together in paralysis as to indicate a close proximity of their respective nuclei.

There have been a number of cases observed, in which the superior and inferior recti were associated together in paralysis, other eye muscles escaping. Thus a case of Wernicke* (12) is recorded, in which after a left hemiplegia there was a disturbance of the movement of both the eyes of such a character, that looking up or down was almost impossible, while lateral motions were well performed, there being at rest an evident paralysis of the left superior rectus (13). There was no ptosis. The autopsy showed an old contracted cicatrix in the right corp. quadrigemina and optic thalamus, which had resulted from a softening. It will be remembered that in the second case of Kahler and Pick (11) there was a paresis of the levator palpebræ rectus superior and inferior, as well as a paralysis of the rectus internus. In the case here presented (14) there was a paralysis of the left superior and inferior recti, no other muscle being affected. And in these cases the obliquus

* Wernicke, Berl. Klin. Woch., 1876, No. 29, and Arch. f. Psych., viii.

DIAGRAM I.—To Show the Associated Paralysis of the Iris Muscles, Levator Palp., and Rectus Superior.

Cases.	Sphincter Iridis and Ciliary Muscle.	Levator Palpebræ.	Rectus Superior.
Mauthner, 27 cases.....		L R	+
Ophthalmoplegia externa totalis.		L R	+
Westphal.....			
Regula.....	L R		
Leube.....	R		
Graefe.....	L R		
Bernhardt, No. 1.....	L R		+
Steffen.....	L R		
Thomsen.....	L R		
Bernhardt, No. 2.....		L R	-
Kahler and Pick, No. 1.....		L	+
Starr, No. 1.....		L	
Lechthelm.....		L R	+
Mauthner.....		L R	
Kahler and Pick, No. 2.....		L	+
..... Paralysis, total or permanent. Paralysis, partial or temporary.	+ Indicates that other muscles were affected.	

inferior was not involved when the superior rectus was affected. It is well known that the action of these two last-named muscles is homologous; but they are not, therefore, necessarily affected together, and cases recorded by Henoch,* (15) by Gowers,† (16) and the case of Thomsen already cited, in all of which the only permanent paralysis was that of the superior rectus in one or both eyes, prove that the obliquus inferior may, and often does, escape when the superior rectus is paralyzed.

DIAGRAM II.—To Show the Associated Paralysis of Rectus Superior and Inferior.

<i>Cases.</i>	<i>Rectus Sup.</i>	<i>Rectus Inf.</i>
Wernicke.....	L —————	—————
	R —————	—————
Starr.....2	L —————	—————
	R —————	————— +
Kahler and Pick..1	L —————	————— +
	R —————	—————
Lichtheim.....	L —————	————— +
	R —————	————— +
Kahler and Pick..2	R —————	————— +
	L —————	—————
Bernhardt.....1	L —————	————— +
	R —————	—————
Parinaud.....8	R —————	—————
	L —————	—————

From these cases we may argue that the centre of the inferior rectus lies next in order to that for the superior rectus. But since in some cases the obliquus inferior has been paralyzed with the superior rectus and without an affection of the inferior rectus, it is necessary to locate its centre equally near to that of the superior rectus. This can

* Henoch, Berl. Klin. Woch., 1864, No. 13.

† Gower's Dis. of Nerv. Syst., 1888, vol. ii., p. 174.

only be done by assuming that the centres for the superior rectus and inferior rectus lie side by side, while that of the inferior oblique lies next to but behind that of the superior rectus.

The last muscle to be considered is the rectus internus.

The fact that the recti interni are concerned in the act of accommodation has led to the supposition that its centre lies in close proximity to the centres for the pupil. And this view has prevailed in spite of the fact that its associated action with that of the abducens of the opposite side would lead to the hypothesis that the centres of the rectus internus and rectus externus would be near together. Paralysis of one rectus alone has not yet been observed, but it has been noted that a paralysis of the act of convergence may occur when the action of the muscles in other movements is preserved, and in six cases published by Parinaud,* the paralysis of convergence was accompanied by defective pupillary action. These cases would indicate that the nucleus of the internal rectus is near to that of the iris. A few cases have been recorded, in which paralysis of the internus has occurred with that of other muscles. Thus is the second case of Kahler and Pick the rectus internus was permanently paralyzed and the levator palpebræ, superior rectus and inferior rectus were paretic. In a case of Lichtheim,† (17) the right rectus internus, levator palpebræ and rectus superior were paralyzed, and the rectus inferior and obliquus inferior were paretic. In a case of Graefe‡ the right rectus internus, levator palpebræ and rectus inferior were paralyzed, (18) and the left rectus internus was weak, while the left levator palpebræ was paralyzed. (19) In my case here published, the right rectus internus was temporarily paretic, and the right rectus inferior was permanently paralyzed. (20) In the cases of Kahler and Pick (No. 1), and Bernhardt (No. 1), the rectus internus was paretic as were also the rectus inferior and levator palpebræ, and rectus superior.

It is evident from these cases that the rectus internus centre is closely related to several other centres, viz., to

* Brain, Oct., 1886.

† Lichtheim, cited by Mauthner. Case 17, l. c.

‡ Graefe, cited by Mauthner. Case 28, l. c.

DIAGRAM III.—To Show Associated Paralysis of the Iris, Ciliary Muscle, Rectus Internus, and Rectus Inferior.

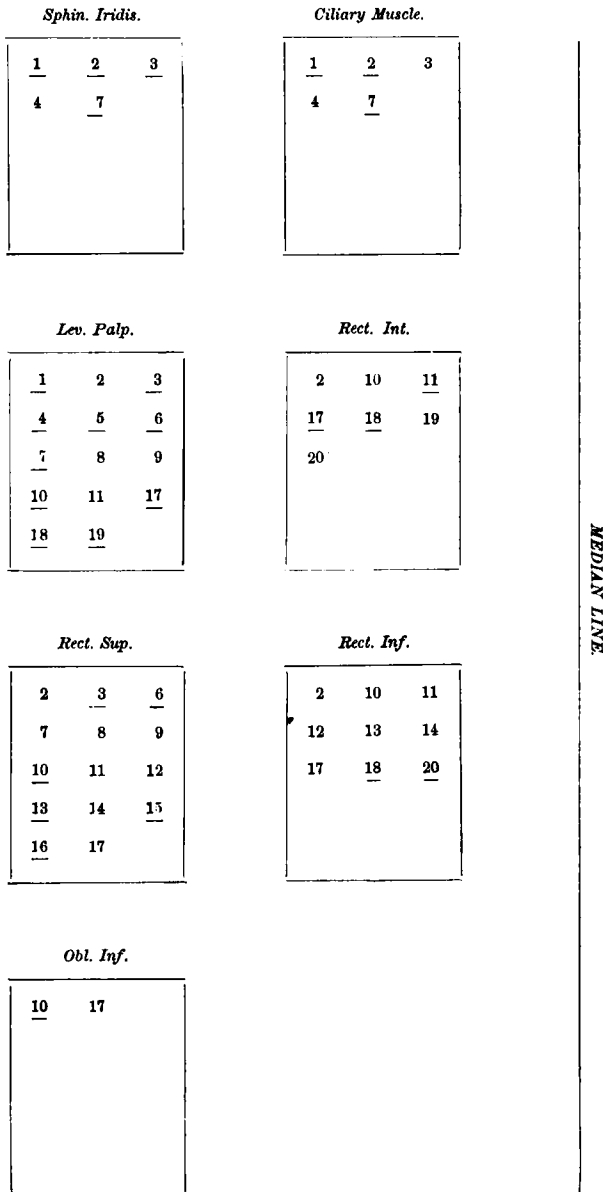
Cases.	Sphincter Iridis.	Ciliary Muscl.	Rectus Internus.	Rectus Inferior.
Parinaud..... 1				
..... 2				
..... 3				
..... 4				
..... 5				
Bernhardt 1				
Mauthner.....				
Graefe.....				
Kahler and Pick..... 2				
Lechthelm.....				
Starr 2				
Kahler and Pick..... 1				

that of the iris, to that of the levator palpebræ, to that of the rectus inferior, and to its fellow on the opposite side. A central position with these muscles on its four sides is therefore the only one which will conform to the facts.

Can any diagram be made out which will explain all these cases. In 1881, an arrangement of the nuclei of the third nerve was proposed by Kahler and Pick, which was based entirely upon the two cases with autopsies here cited. And it must be confessed that when those autopsies are studied it seems as if the conclusion drawn was hardly warranted by the premises; for the lesions in each case involved the nerve roots of the third nerve rather than their nuclei; and this has been the case in most of the cases of partial ophthalmoplegia externa hitherto published. But the arrangement proposed by these authors has shown itself competent to explain the various cases as they have arisen since 1881, and the cases here brought together afford a brilliant confirmation of the essential part of the diagram of Kahler and Pick, and establish it, I believe, for the first time, upon a sufficient basis. It is true that Mauthner, in his lectures, has approved of Kahler and Pick's arrangement in preference to that of Hensen and Völchers', but he cites but five cases, and admits that the evidence is scanty. The cases here collected are twenty in number, each eye being counted as one case, unless both eyes were affected alike. The following diagram shows the relative position of the nuclei, and the extent and degree of the paralysis in each case cited, the numbers corresponding to the cases numbered in the text, and being underlined when the paralysis was complete or permanent. For convenience, the arrangement on the left side of the raphé only is represented, all the cases being tabulated as if on the left side.

The relative position of the centres for the sphincter iridis and ciliary muscle is still an open question. The observation of Westphal is sufficient to indicate that they lie side by side. Kahler and Pick placed the centre for accommodation in front of that for the light reflex. But the observation of Parinaud cited would indicate that the centre for accommodation being usually affected with that of the

DIAGRAM IV.—*The Position of the Third Nerve Nuclei
on the Left Side.*



rectus internus lies nearer to it than does the centre for the iris. The number of fibres passing from the corpora quadrigemina to the lateral of the two nuclei described by Westphal seems greater than the number passing to the median nucleus, and as these fibres probably complete the reflex arc for light reflex joining the optic and iris centres, I have placed the sphincter iridis nucleus outside of that for the ciliary muscle. It will be noticed that this arrangement brings the ciliary muscle centre near to that of the rectus internus as the cases of Parinaud require. Another fact which can be deduced from the diagram is the relative frequency of affection of the various muscles, it being evident that the superior rectus and levator palpebræ are most frequently paralyzed, and the superior oblique is rarely involved.

One pathological fact of importance remains to be noticed. In all the cases recently examined after death, in which one or more of the muscles of the third nerve was paralyzed, but in which total ophthalmoplegia externa was not present, the lesion found did not involve the nuclei of the third nerve in the aqueduct of Sylvius, but the roots of the nerve on their way from those nuclei through the tegmentum to their exit from the crus. These roots pass through the red nuclei of the tegmentum. These nuclei are closely connected with the superior peduncles of the cerebellum. It is not at all unlikely, therefore, that the vertigo which so frequently occurs in such cases is due not so much to the disturbance of vision, but to the lesion of these nuclei, and is comparable to the vertigo of cerebellar disease. In my own cases the patients complained of vertigo in the dark and when the eyes were closed, as well as when they were open. It seems warrantable, therefore, to conclude that in any case of partial ophthalmoplegia externa associated with vertigo the diagnosis of a lesion in the tegmentum cruris affecting the red nuclei may be made, provided the vertigo is not ocular in origin. While in cases of total ophthalmoplegia externa in which vertigo is not a marked symptom the diagnosis of a lesion in the gray matter of the aqueduct of Sylvius above the tegmentum is justifiable.

CONCLUSIONS.

Therefore, in any case in which the muscles moving the eyeball are involved, it may be possible to locate the lesion. If the iris alone is affected, the lesion is small, and lies either in the ciliary ganglion in the orbit, or just at the opening of the aqueduct of Sylvius into the third ventricle. If all the muscles of the eyeball are affected together, the external rectus and superior obliquus, as well as those supplied by the third nerve, *excluding the iris*, the case is one of ophthalmoplegia externa totalis, and the lesion lies in the gray matter of the floor of the fourth ventricle and of the aqueduct of Sylvius. Both eyes are then involved. If all the muscles of the eyeball supplied by the third nerve are affected, *including the iris*, the case is one of total peripheral paralysis of the third nerve, and the lesion lies on the base of the brain, and may in time implicate other cranial nerves. One eye is usually alone affected.

If one or two of the muscles of the eyeball supplied by the third nerve are affected, others escaping, the lesion lies in the tegmentum of the crus cerebri, between the nuclei of origin and the point of exit of the third nerve. One eye or both may be affected, but both eyes are rarely affected in the same manner.

There is but one exception to the last conclusion, and that is in the case of post diphtheritic ocular paralysis, in which the peripheral branches of the third nerve are affected after the entrance of the nerve trunk into the orbit. And here the existence of a diphtheria preceding the paralysis will establish the diagnosis.